Juvenile Nasopharyngeal Angiofibroma: Imaging Characteristics and Pre-Operative Embolization

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Juvenile nasopharyngeal angiofibroma (JNA) is a benign highly vascular tumor. It almost exclusively affects male adolescents and young adults. Nasal blockage and recurrent epistaxis are the classic symptoms. Cross–sectional imaging is vital for accurate diagnosis and operative planning. Pre-operative embolization is frequently performed to decrease blood loss and has especially facilitated endoscopic resection.

A sixteen-year-old male presented with one-month complaint of unilateral nasal blockage and frequent epistaxis. Angiofibroma was diagnosed by CT and MRI. Angiography followed by embolization was performed 24 hours prior to endoscopic surgical resection. The reported blood loss was less than 50 ml. No complications were encountered.

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JNA is a rare vascular tumor that accounts for 0.05% to 0.5% of all head and neck neoplasms; however, it is the most common tumor of the nasopharynx in young males[1]. The etiology of JNA remains unknown; however, the association with androgen receptors, chromosomal alterations and growth factors has been described[2-4].

The term angiofibroma was first used by Friedberg in 1940. The name indicates fibrous and vascular components, which of these is the origin, is a matter of debate[5]. CT and MRI imaging could achieve the diagnosis.

The aim of this presentation is to report a case of Juvenile Nasopharyngeal Angiofibroma, which was managed by super-selective embolization prior to endoscopic surgical resection.

THE CASE

A sixteen-year-old male presented with a one-month complaint of unilateral nasal blockage and frequent epistaxis. No other physical or laboratory abnormality detected. Plain paranasal-sinuses and CT revealed left-sided posterior nasal polypoidal mucosal thickening occluding the left choanae and extending posteriorly through nasopharyngeal cavity and widening of left sphenopalatine foramen with bowing of posterior maxillary wall "Holman-Miller sign". Appreciable infiltration of the medial wall of the base of pterygoid plates was found, see figure 1.

Enhanced MRI revealed a posterior nasopharyngeal mass measuring 3.2 cm x 2.2 cm, appearing iso intense on T1 weighed images and of heterogeneous intensity at T2WI with subtle hyperintense foci, and thin flow voids visible on T2 weighed images, see figure 1.

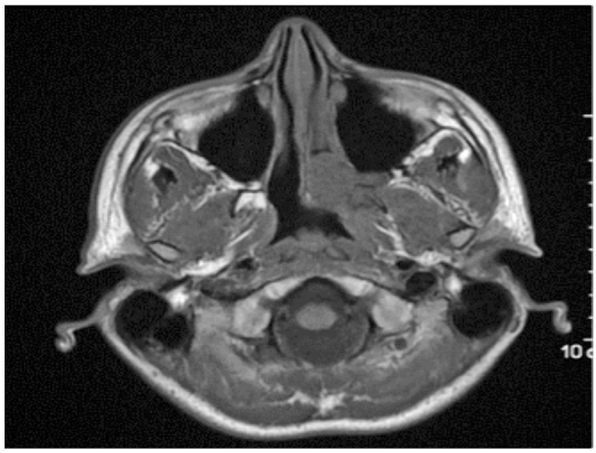

Figure 1: Contrast Enhanced Paranasal-Sinuses MRI T1WI Revealed a Posterior Nasopharyngeal Mass Isointense Lesion. The Mass Was Seen Originating from Sphenopalatine Foramen

The mass was seen originating from sphenopalatine foramen causing its subsequent widening. It extends medially into the nasopharynx, laterally through pterygopalatine fossa. Pterygomaxillary fissure was widened, and the mass is infiltrating the base of pterygoid plates and extended anteriorly. Bowing of the posterior wall of maxillary antrum anteriorly "Holman-Miller sign" was found. No intracranial or cavernous sinus invasion found, as well as no sphenoidal sinus invasion, see figure 2. The mass showed evident post-gadolinium enhancement suggestive of hypervascularity. Angiography followed by embolization was performed 24 hours prior to endoscopic surgical resection. The reported blood loss was less than 50 ml.

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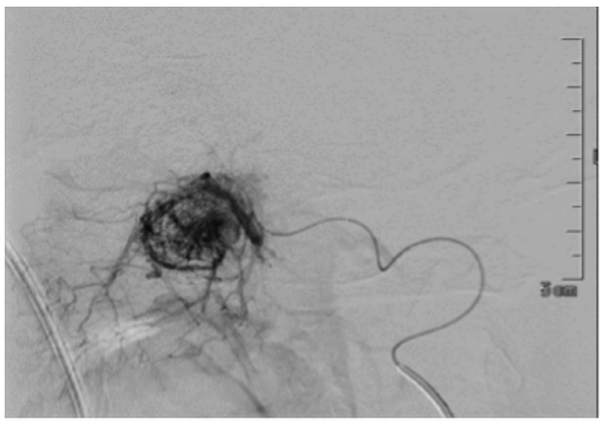

Figure 2: Super Selective Coaxial Angiogram of Distal Left Internal Maxillary Artery

No complications were encountered, and the patient was discharged home on the third day.

The procedure initiated by securing the right common femoral artery with 5 F sheaths, through which 5 F Win catheter was advanced through right common iliac artery to the abdominal aorta, the catheter was advanced to the aortic arch followed by arch aortogram delineating the arch vessels. Cannulation of left CCA was performed followed by carotid angiogram showing ICA and ECA as well as the blush of the nasopharyngeal angiofibroma. Selective cannulation of left ECA using a coaxial micro catheter over micro wire was achieved. Angiogram revealed the feeding vessels of the tumor which originated mainly from the left internal maxillary artery.

Embolization of the feeding arteries was performed using diluted tri-acryl gelatin microspheres 300-500 µm under fluoroscopy, and continued until complete stasis of the flow at feeding artery was achieved with total absence of tumor blush; furthermore, the main feeder artery was embolized by using a 3 mm x 6mm 0.010 HydroFill Endovascular Embolizing coil.

Post procedure angiogram revealed successful embolization with significant tumor blush reduction, see figure 3.

Figure 3: Post-Embolization Angiogram Showed Significant Reduction in Blush

Femoral access sheath was removed and hemostasis was secured by manual compression and pressure bandage.

No complications were encountered during or immediately after the procedure.

DISCUSSION

Although histologically benign, JNA could display aggressive behavior by causing local destruction and invasion. Typically, males between 15-29 years of age and present with a history of nasal blockage and epistaxis. Nasal speech, proptosis, facial asymmetry and many others could occur with advanced disease[6]. The tumor is thought to originate from the sphenopalatine fossa, and it could extend to any direction[6,7]. Skull base and intracranial invasions at presentation are not rare[8]. JNA takes the pathway of highest resistance to reach its destination[7]. Diagnostic imaging is the corner stone for evaluation and treatment of JNA[9]. CT is important for delineating bony erosions and remodeling, which is used for staging of the tumor[9-11].

MRI is superior to CT in detecting intracranial extension. JNA appears as hypo or iso-intense mass in T1 or heterogonous on T2 weighted images due to the fibrous component, but show intense enhancement with gadolinium[10].

Angiography in early arterial phase would reveal dense blush consistent with the venous phase[9]. JNA blood supply is from the ipsilateral external carotid artery and usually from internal maxillary artery branches, also could be from ascending pharyngeal and palatine arteries[11]. Bilateral blood supply from both external carotid systems was reported[12].

Embolization is an urgent procedure to stop an intractable hemorrhage or commonly elective in the preoperative stage. The definite treatment is by open surgical or endoscopic approach. Adjunct preoperative embolization is intended to decrease blood loss, operative time and reduce the risk of recurrence[13].

The most dreaded complication (although rare) is the migration of embolizing material to the intracranial circulation which has grave neurological sequelae; it could lead to blindness, internal carotid stroke, facial necrosis, cranial nerve palsies and vertebrobasilar stroke[14,15]. Minor complications include short-term pain, headache and facial edema[15]. Embolization must be performed 24 to 48 hours prior to surgical resection to avoid revascularization[16].

Most studies found a significant reduction in blood loss in pre embolized cases, but some found no advantage[17-22]. In addition, some studies found that embolization makes tumor resection difficult by obscuring the tumor borders[23,24].

Percutaneous embolization is an alternative to the classic transarterial approach. Some studies have successfully used Onyx[25,26]. In addition, a hybrid approach has been described[27].

CONCLUSION

A mass in the sphenopalatine fossa that enhances with contrast on CT or MRI in the appropriate age group should raise the suspicion of JNA. Interventional radiology has a

essential role in diagnosis and management of this tumor. Careful pre-operative embolization is a safe procedure which could decrease intraoperative blood loss.

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